DISEASES OF THE RESPIRATORY SYSTEM 2017

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LECTURE 2: ATELECTASIS AND EMPHYSEMA

INTRODUCTION

In this lecture we will discuss atelectasis which is a complication of several medical and surgical conditions

Then we will talk about obstructive and restrictive lung diseases and the difference between them. And we will discuss the first (out of four) obstructive lung disease which is emphysema.

Reference:

Robbins 9th edition: atelectasis: page 460, emphysema: 462-466

10th: atelectasis 495-496, emphysema: 498- 501

ATELECTASIS =انْخِماص

Atelectasis refers to the collapse of inflated lung, producing areas of relatively airless pulmonary parenchyma. The main types are:

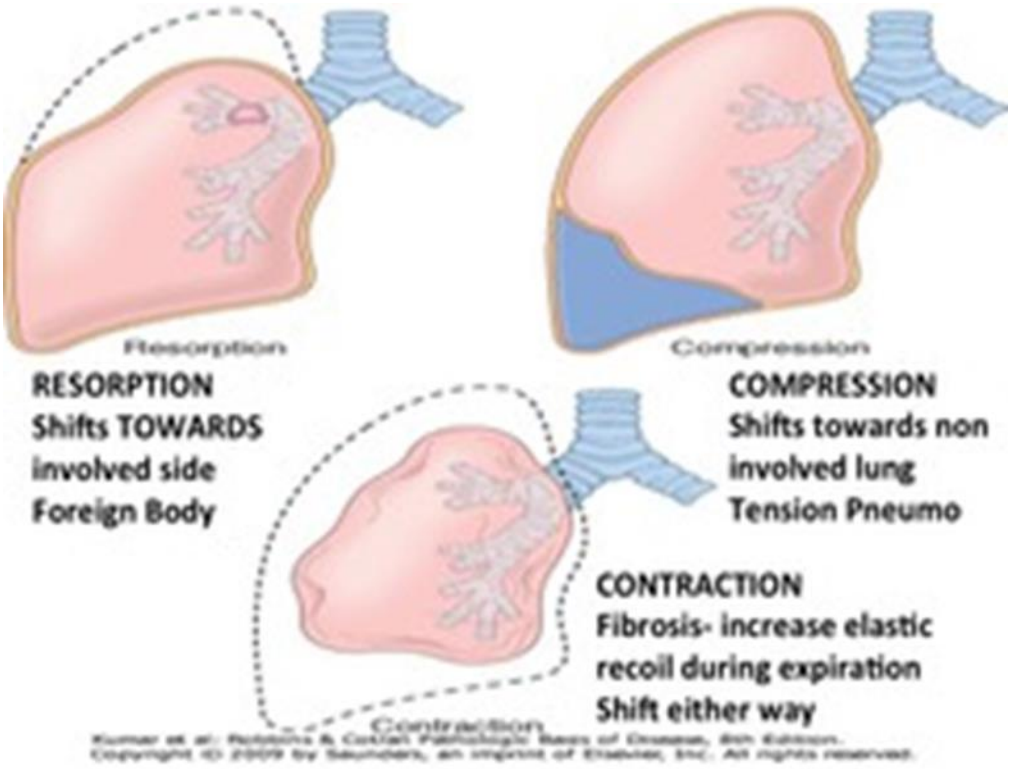
• **Resorption atelectasis** stems from complete *obstruction* of an airway. Over time, air is resorbed from the dependent alveoli, which collapse. Since lung volume is diminished, the mediastinum shifts toward the atelectatic lung.

Airway obstruction is most often caused by excessive secretions (e.g., mucus plugs) or exudates within smaller bronchi, as may occur in bronchial asthma, chronic bronchitis, bronchiectasis, and postoperative states. Aspiration of foreign bodies and, rarely, fragments of bronchial tumors may also lead to airway obstruction and atelectasis.

• **Compression atelectasis** results whenever significant volumes of fluid (transudate, exudate or blood), tumor, or air (pneumothorax) accumulate within the pleural cavity. With compression atelectasis, the mediastinum shifts away from the affected lung.

So any accumulation of material within the pleural space causes compression atelectasis: -Fluid (pleural effusion) -Blood (haemothorax) -Air (pneumothorax)

• **Contraction atelectasis** occurs when focal or generalized pulmonary or pleural fibrosis prevents full lung expansion.



Atelectasis (except when caused by contraction) is potentially reversible and should be treated promptly to prevent potential complications like: hypoxemia and superimposed infection of the collapsed lung.

TREATMENT

Treatment is directed at correcting the underlying cause. Post-surgical atelectasis is treated by physiotherapy, focusing on deep breathing and encouraging coughing. An incentive spirometer (pic below) is often used as part of the breathing exercises. Walking is also highly encouraged to improve lung inflation.

OBSTRUCTIVE VERSUS RESTRICTIVE LUNG DISEASES

Chronic, primary lung diseases are usually divided into obstructive and restrictive diseases.

Obstructive lung diseases (or obstructive airway diseases) are characterized by an increase in resistance to airflow due to partial or complete obstruction at any level from the trachea and larger bronchi to the terminal and respiratory bronchioles.

So in obstructive lung diseases, there is obstruction of the airway which could be due to **structural** damage with associated mucin secretion (like in chronic bronchitis) or due to **functional** obstruction where there is no actual physical obstruction but the function of the alveoli is affected by being dilated, like in emphysema.

In both structural and functional obstruction, there is a **relatively normal air entry** into lungs (inspiration is normal) because inspiration is a passive process. But expiration is affected because you need effort to expire (expiration is an active process that needs muscle contraction). This means air entry is normal (inspiratory volumes normal) **and lung capacity is normal** , or slightly decreased (there is no problem in how much air the lungs can get, this is measured by the forced vital capacity (FVC).

FVC= is the maximum amount of air a person can exhale from the lungs after a maximum inhalation. It is equal to the sum of inspiratory reserve volume, tidal volume, and expiratory reserve volume. This volume is normal in people with obstructive lung diseases.

We said these patients have a problem in expiration, we measure expiration by the **FEV1= forced expiratory volume in the first second**, this is the volume of air that can forcibly be exhaled in the first second, after maximum forced inspiration. In obstructive diseases this volume is decreased

So the ratio between FEV1 / FVC is reduced. This is the most important test to differentiate obstructive from restrictive lung diseases. *An FEV1/FVC ratio of less than 70% generally indicates airway obstruction* (please note some references quote different number, don't worry about numbers, just get the idea)

Restrictive diseases are characterized by **reduced expansion** of lung parenchyma **and decreased total lung capacity.** In restrictive diseases are associated with proportionate decreases in both total lung capacity and FEV1, leading to normal FEV1/FVC ratio. In healthy adults this should be approximately 70–85%

OBSTRUCTIVE LUNG DISEASE AE MAINLY FOUR DISEASES:

1. emphysema
2. chronic bronchitis
3. bronchial asthmaa
4. bronchiectasis

NOTE: Emphysema and chronic bronchitis are called chronic obstructive pulmonary disease (COPD)

Obstructive vs restrictive lung diseases

Obstructive	restrictive
characterized by limitation of airflow due to partial or complete obstruction	characterized by reduced expansion of lung parenchyma accompanied by decreased total lung capacity.
Eg are emphysema, chronic bronchitis, bronchiectasis, and asthma	Eg are ILD like Fibrosing alveolitis, idiopathic pulmonary fibrosis, interstitial pneumonia, Pneumoconiosis,Sarcoidosis; and chest wall neuromuscular diseases
total lung capacity normal	decreased
forced vital capacity (FVC) normal	reduced
decreased expiratory flow rate, measuerd as forced expiratory volume at 1 second (FEV_1)	Normal or reduced
FEV1/FVC ratio < 0.80	normal

OBSTRUCTIVE VS. RESTRICTIVE

Obstructive disorders	Restrictive disorders
• **Characterized by:** reduction in airflow. • So, shortness of breath → in exhaling air. (the air will remain inside the lung after full expiration) 1. COPD 2. Asthma 3. Bronchiectasis	• **Characterized by** a reduction in lung volume. • So, Difficulty in taking air inside the lung. (DUE TO stiffness inside the lung tissue or chest wall cavity) 1. Interstitial lung disease. 2. Scoliosis 3. Neuromuscular cause 4. Marked obesity

EMPHYSEMA = نفاخ رئوي

As we mentioned above, chronic bronchitis and emphysema are called COPD or COAD (chronic obstructive airway disease). These two entities are distinct diseases but they **overlap** and **coexist** in the same patient because both are caused by smoking. Their pathogenesis, definition and histological features are different.

One of the main differences between these two diseases is that the definition of emphysema is morphologic (depends on histological and macroscopic appearance) , whereas chronic bronchitis is defined on the basis of clinical features.

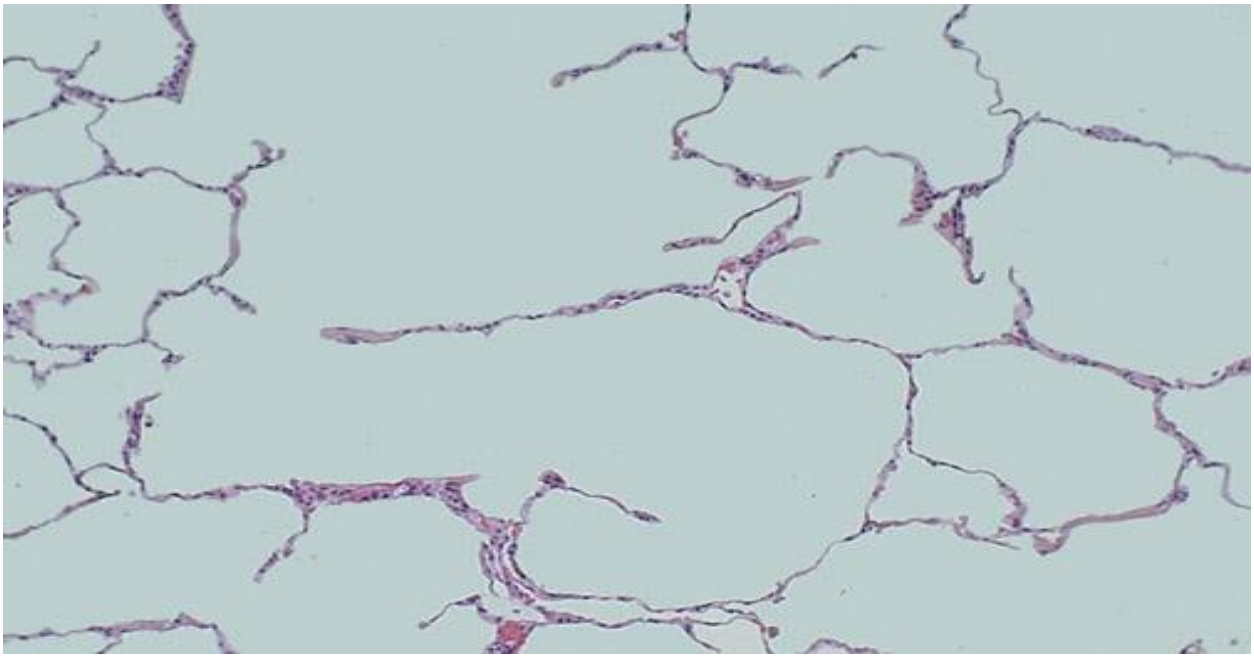
The anatomic distribution is also different; chronic bronchitis initially involves the large airways, whereas emphysema affects the acini.

Clinical presentation is also differs (details later)

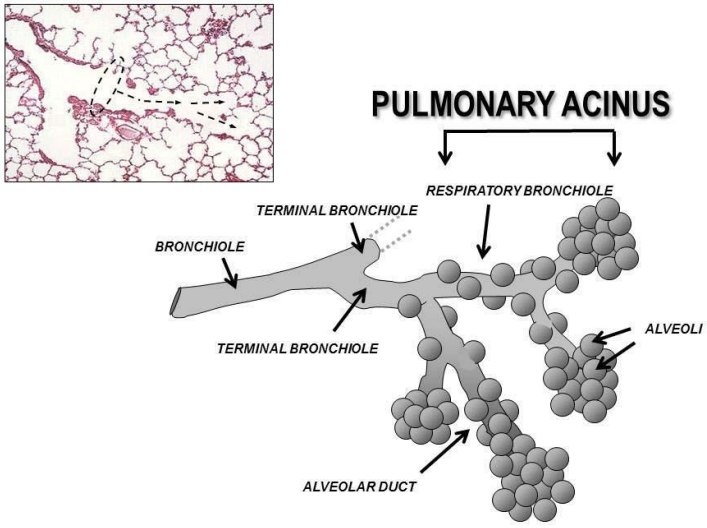
DEINITION OF MPHYSEMA:

Abnormal permanent enlargement of the air spaces distal to the terminal bronchioles accompanied by destruction of their walls **without fibrosis**

Note in the pic below how the alveoli are enlarged, the walls between them disappeared causing this enlargement. Note also that there is no fibrosis.



Emphysema is classified according to its anatomic distribution within the lobules and acini . An acinus is the structure distal to terminal bronchioles, and a cluster of three to five acini is called a lobule



There are four major types of emphysema according to distribution in relation to acini: Centriacinar, Panacinar, Distal acinar, and irregular.

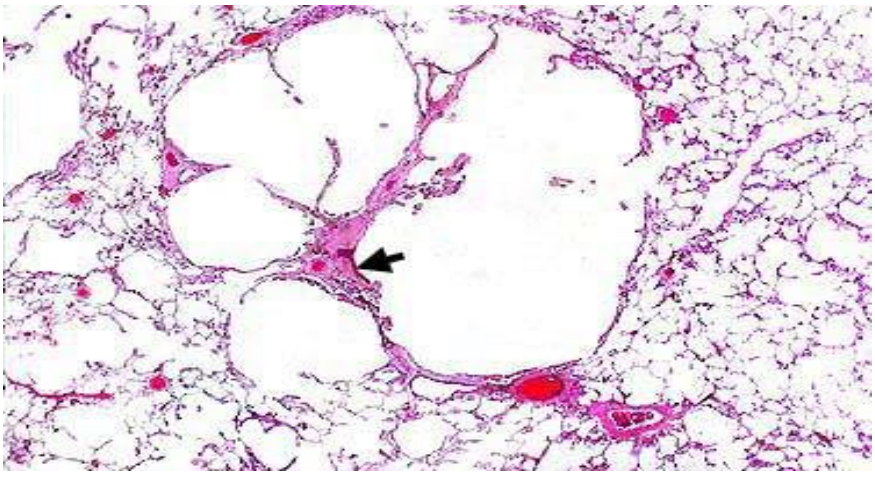
1. **<u>centriacinar, or centrilobular emphysema:</u>**

In this type the central or proximal parts of the acini, formed by respiratory bronchioles, are affected, while distal alveoli are spared . So both emphysematous and normal air spaces exist within the same acinus and lobule

In this type the dilated respiratory bronchioles are more common and severe in the **upper lobes.**

-This type of emphysema is most commonly caused by smoking.

Centriacinar emphysema: note that there are dilated alveoli and normal ones.



2. **Panacinar (Panlobular) Emphysema**

In this form the acini are **uniformly enlarged**, from the level of the respiratory bronchiole to the terminal blind alveoli .It tends to occur more commonly in the lower lung zones and It occurs in α1-antitrypsin deficiency.

So: centriacinar is commoner than panacinar, because number of people who smoke is more than those who have alpha 1 antitrypsin deficiency!

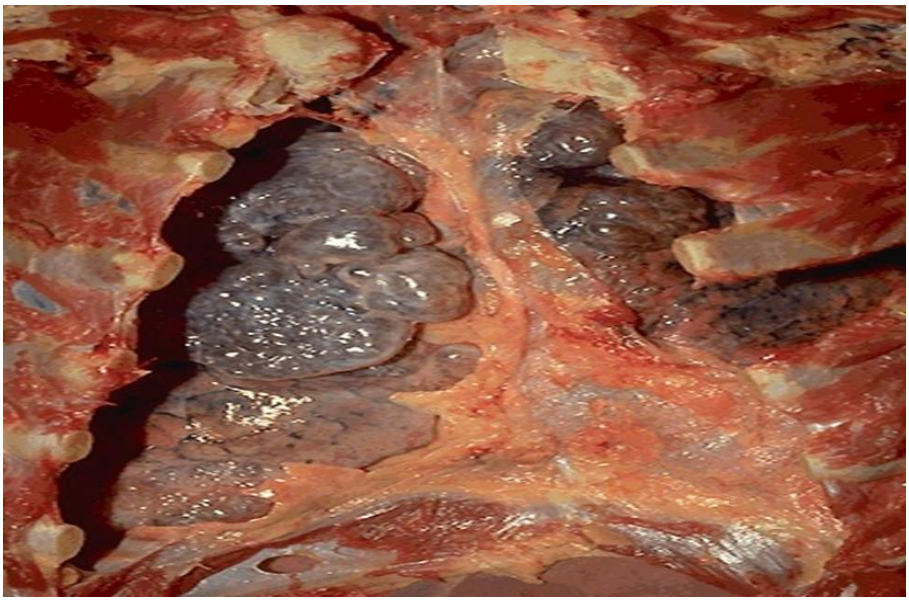
3. **Distal Acinar (Paraseptal)**

It involves the distal part of the acinus. The proximal portion of the acinus is normal

This form is more striking adjacent to the pleura, along the lobular connective tissue septa.

The characteristic finding is the presence of multiple, enlarged air spaces ranging in diameter from less than 0.5 mm to more than2.0 cm, sometimes forming cystic structures that, with progressive enlargement, are referred to as bullae

Ruptured bullae cause spontaneous pneumothorax especially in young adults. Bullae seen in pic below:



4. **Irregular Emphysema**

The acinus is irregularly involved. This form is almost invariably associated with scarring, such as that resulting from healed inflammatory diseases. Although clinically asymptomatic, this may be the *most common form of emphysema.*

PATHOGENESIS

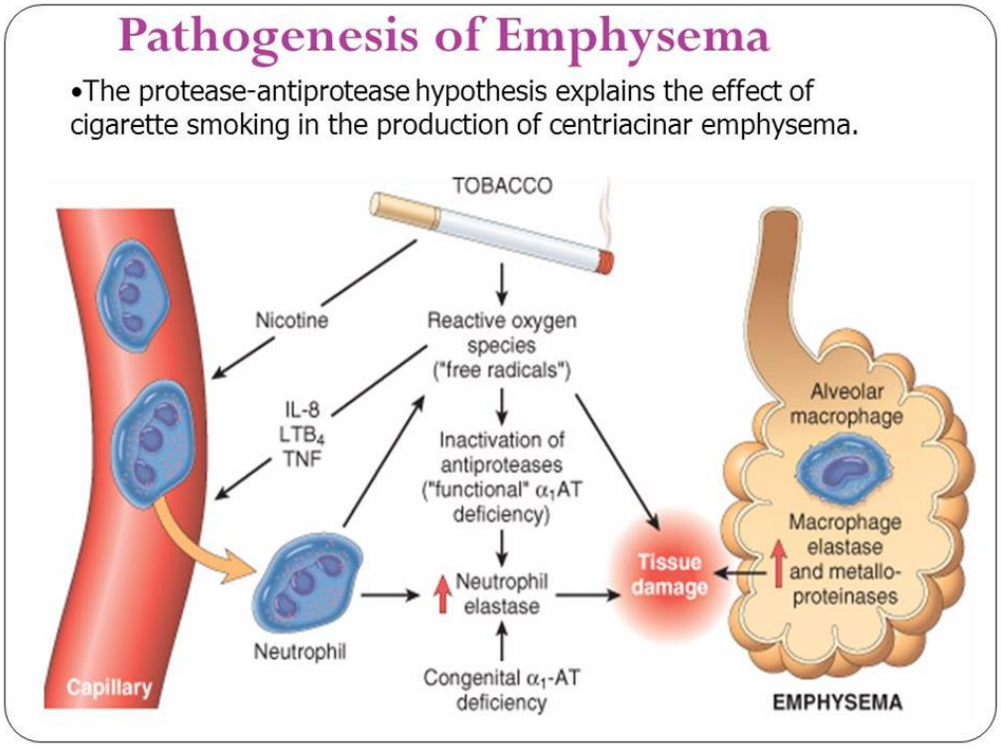
Emphysema is caused by exposure to toxic substances such as tobacco smoke and inhaled pollutants which induce inflammation with accumulation of neutrophils, macrophages and lymphocytes in the lung.

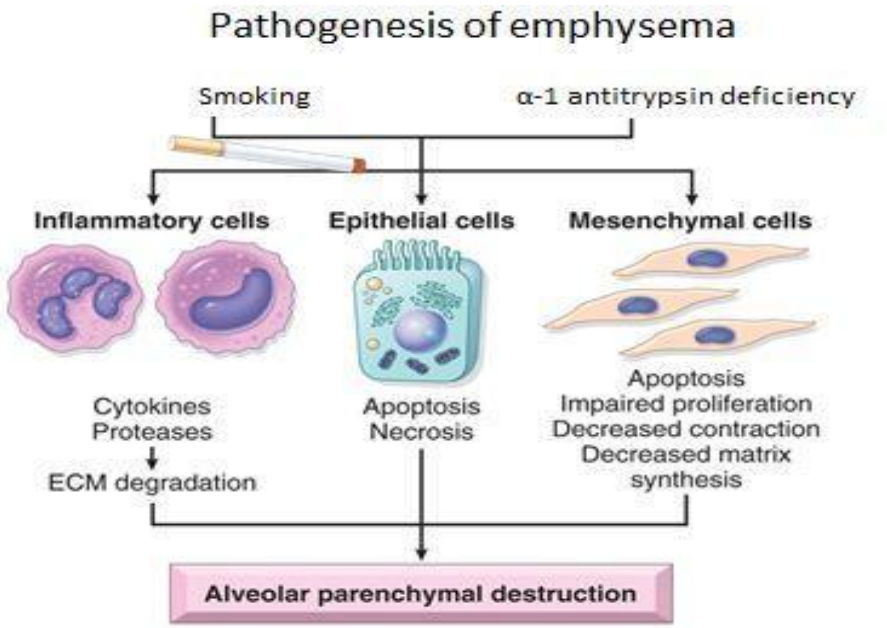
Neutrophils release elastases, cytokines (including IL-8) and reactive oxygen species causing epithelial injury and proteolysis of the extracellular matrix (ECM). Unless checked by antielastases (e.g., α1-antitrypsin) and antioxidants, the cycle of inflammation and ECM proteolysis continues.

More than 80% of patients with congenital α1-antitrypsin deficiency develop symptomatic panacinar emphysema, which occurs at an earlier age and with greater severity if the affected person smokes.

Note that in emphysema there is loss of not only epithelial and endothelial cells but also mesenchymal cells, leading to lack of extracellular matrix, the scaffolding upon which epithelial cells would have grown. That's why the destruction is not accompanied by fibrosis. Thus, emphysema can be thought of as resulting from insufficient wound repair.

With the loss of elastic tissue in the surrounding alveolar septa, radial traction on the small airways is reduced and as a result, they tend to collapse during expiration-an important cause of chronic airflow obstruction in severe emphysema





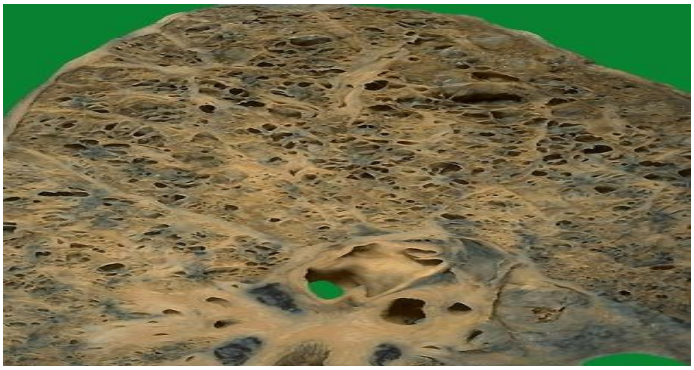
GENETIC FACTORS

Multiple genetic factors control the response to injury after smoking. The TGFB gene exhibits polymorphisms that influence susceptibility to the development of COPD by regulating the response of mesenchymal cells to injury. Metalloproteinases play a role: MMP-9 gene polymorphisms and higher levels of both MMP-9 and MMP-12 have been found in some emphysema patients. Moreover, MMP-12-deficient mice are protected from cigarette smoke-induced emphysema.

With certain polymorphisms, mesenchymal cell response to TGF-β signaling is reduced, which results in inadequate repair of elastin injury caused by inhaled toxins

MORPHOLOGY

Lungs are spongy. Histologic examination reveals :
a. Destruction of alveolar walls without fibrosis, leading to enlarged air spaces
b. The number of alveolar capillaries is diminished.



CLINICAL FEATURES

Dyspnea usually is the first symptom which begins insidiously but is steadily progressive. Weight loss is common and may be so severe as to suggest a hidden malignant tumor. The cause of weight loss is not exactly known but could be due to dryness of the mouth as patients breathe from the mouth, this dryness causes decreased sensation of taste buds and decreased appetite.

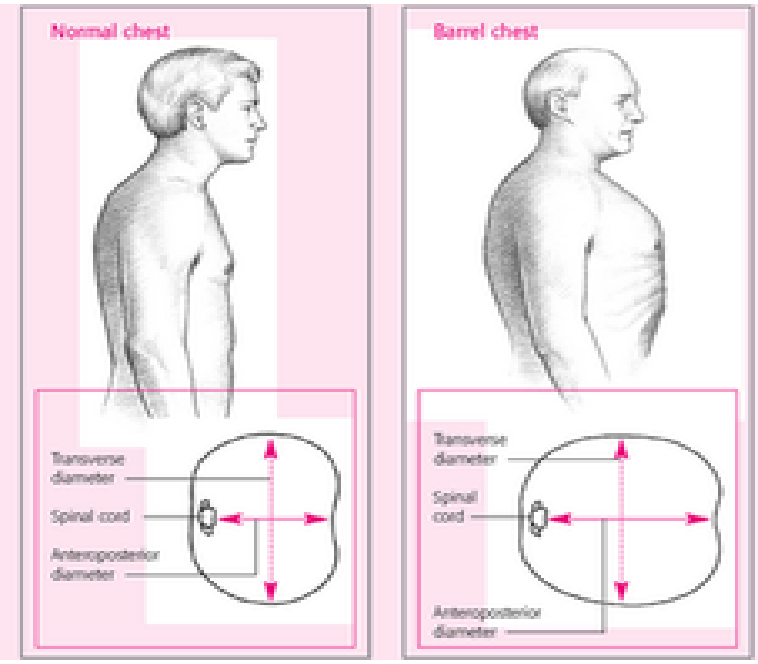
If a patient has pure emphysema and no element of chronic bronchitis he will have:

a. Barrel-chest, b. with obviously prolonged expiration, c. sitting forward in a hunched-over position, attempting to squeeze the air out of the lungs with each expiratory effort (they try to use their muscles to squeeze air out during expiration)

Barrel chest = بَرْميل , refers to an increase in the *anterior posterior diameter of the chest wall* resembling the shape of a barrel, most often associated with emphysema. There are two main causes of the barrel chest phenomenon in emphysema:

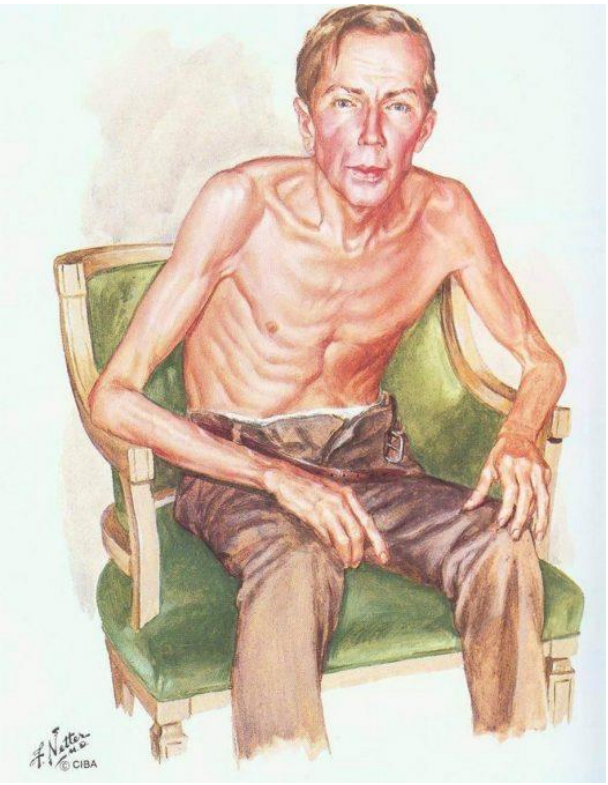
1. Increased compliance of the lungs leads to the accumulation of air pockets inside the thoracic cavity.

2 .Increased compliance of the lungs increases the intrathoracic pressure. This increase in pressure allows the chest wall to naturally expand outward



Dyspnea and hyperventilation are prominent, so that until very late in the disease, gas exchange is adequate and **blood gas values are relatively normal.** The obstruction in these patients is functional, there is no anatomic or structural obstruction. They have problem with expiration because they lost the elastic fiber around the alveoli needed to squeeze the alveoli to get rid f the air in them during expiration. So these patients try to overcome this problem by acquiring a position (the hunched position) to use their muscles and rib cage to squeeze the air out of the alveoli. They labor for expiration and their expiration is prolonged, as if they puff it. This puffing helps getting rid of the CO2 and the patients continue having normal O2 and CO2 levels (no cyanosis, they stay "red or pink "). Because of this red color and the puffing described above, these patients who suffer pure emphysema are described as pink buffers

Pink buffer: note the skin color, the position, the weight loss



<u>Complications of emphysema:</u>

Secondary pulmonary hypertension
Right sided heart failure
Respiratory failure

THANK YOU